Cardiac resynchronization therapy (CRT) for patients with congenital heart disease is a promising therapeutic option as shown in the report by Sakaguchi et al. in this issue of the Journal. In their study, the clinical response to CRT was compared among systolic ventricular anatomical morphologies: systemic left ventricle, systolic right ventricle, and single ventricle (Figure 1). The results from the single center, which is one of the most experienced, high-volume institutes in Japan, showed CRT benefit in patients with systemic left or single ventricular morphology. However, 7 patients with systemic right ventricle (RV) did not achieve apparent 6-month response from CRT, showing no effect on

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**Figure 1.** Congenital heart disease anatomical subgroups after surgical repair. (A) Example of a syst LV. Repair of tetralogy of Fallot with patch closure of a ventricular septal defect and a right ventricular (RV) outflow/main PA outflow patch. The left ventricle is the systemic ventricle. The right ventricle is dilated because of pulmonary regurgitation. In tetralogy of Fallot, systemic left ventricular failure may develop. (B) Example of syst RV. Venous switch echocardiogram shows the apical 4-chamber view after the Senning procedure in a patient with transposition of great arteries. Mustard repair is another atrial switch operation explained in the schematic. The morphological right ventricle is the systemic ventricle in patients with transposition of great arteries undergoing an atrial switch operation. Systemic RV failure may develop in one-third of cases of congenital/surgical corrected transposition of great arteries. (C) Example of a single ventricle. Apical view of RV morphology with hypoplastic left ventricle. The diagram shows an example of Fontan repair for single ventricular morphology, right atrial to PA anastomosis with atrial “baffle” and ligation of proximal PA for ventricular inversion, transposition of great arteries, double-inlet ventricle, and hypoplastic subaortic outlet chamber. The failure of the single ventricle is common in patients with a hypoplastic left ventricle. IVC, inferior vena cava; PA, pulmonary artery; SVC, superior vena cava; syst LV, systemic left ventricle; syst RV, systemic right ventricle.
either the mean right ventricular ejection fraction (RVEF) or end-diastolic volume index.

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**CRT for the Failing Systemic RV**

There have been mixed results of CRT for patients with a systemic RV. According to 3 multicenter retrospective cohorts, response defined as NYHA improvement ≥1 class or RVEF improvement was observed in only 2 of 9 patients (22%),19 of 27 patients (79%), and NYHA improvement was observed in 13 of 17 patients (76%). Even in the study reporting a relatively good response, the response rate was significantly less than that of systemic left ventricle (LV) patients. Therefore, the study results imply that a detailed evaluation prior to CRT implantation may be crucial to identify who will benefit from CRT, especially for patients with systemic RVs.

**Assessment of RV Electrical Dyssynchrony**

There is currently no consensus for CRT indication in congenital heart disease patients. Adult heart failure guidelines are based on evidence from prospective randomized control trials including several thousands of participants. According to those guidelines, CRT is recommended as class I with level of evidence A, if the patient has LV ejection fraction ≤35%, QRS duration ≥150 ms with left bundle branch block morphology, and NYHA class III–IV despite optimal medical therapy. In the adult population, right bundle branch block (RBBB) morphology (with systemic LV) is generally thought to confer smaller benefit of CRT. There has been no consensus on whether the CRT recommendation applies to systemic RV and wide QRS duration with a narrow QRS (<130 ms), even when there is mechanical dyssynchrony. Therefore, the presence of electrical delay must be the essential requirement for CRT to be beneficial. Three-dimensional electrical activation contact mapping of the RV has reported the RBBB results in RV electrical dyssynchrony and may provide detailed understanding of systemic RV electrical propagation.

**Assessment of RV Mechanical Dyssynchrony**

As mentioned, only prolonged QRS duration and the ECG morphology are the principles defining the CRT guideline in adult cardiology, because mechanical dyssynchrony assessed by conventional echocardiography has been concluded “as useless” by a multicenter trial. However, in the complex anatomy in congenital heart disease, surface ECG findings might not be accurate enough to identify CRT responders. Thus, cardiac imaging of mechanical dyssynchrony may improve selection of congenital heart disease patients who will benefit from CRT. Echocardiography is the most promising noninvasive imaging modality to evaluate mechanical dyssynchrony. Eye replacement of the presence of the septal flash or
shuffle motion has been reported as a simple technique and strongly predictive for CRT response; however, Sakaguchi et al. confirmed this movement in all patients characterized as non-responder. Recently, 2D and 3D strain echocardiography has been focused on as an accurate method of assessing mechanical dyssynchrony. Based on the available evidence, a flow chart indicating the possible CRT strategy for systemic RV is proposed in Figure 2.

Conclusions

New cardiac imaging modalities for assessment of electrical and mechanical dyssynchrony are required to provide more detailed information on the CRT indication in each patient with systemic RV failure.

References